

peer reviewed CASE REPORT

A case report of histiocytic sarcoma of the hepatic flexure

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Abstract

This case report discusses a female patient who presented with a large palpable mass in her right upper abdominal quadrant. A diagnosis of histiocytic sarcoma was made. Her clinical history and management as well as the aetiology, epidemiology and histo-pathological appearances of this disease are discussed.

Keywords

Colonic mass, lymph nodes, spiculated lesions, eosinophilic cytoplasm.

Case report

A female patient in her mid-50s presented with a large palpable mass in her right upper abdominal quadrant. She had no previous history of surgery. She complained of a swollen abdomen for four months before seeking medical treatment. She also experienced night sweats and loss of weight and appetite. She experienced melena for a year before admission. She did consume alcohol and was a known smoker.

She underwent an abdominal ultrasound at a secondary hospital and this examination revealed large abdominal nodes within the abdomen. She was referred to an academic hospital for further management. She underwent computed tomography (CT) scans of the chest and abdomen. An enhanced arterial scan with

a 25 second delay and portal venous scan with a 70 second delay was performed.

The chest CT showed left and right apical spiculated lesions. In addition there were several more spiculated lung lesions and mediastinal nodes. There were multiple enlarged lymph nodes within the abdomen. The CT scan of the abdomen revealed an intussusception of the bowel (Figure 1) and a large colonic mass located at the hepatic flexure (Figure 2).

The patient was admitted to the colorectal unit. A colonoscopy was performed which revealed a large, obstructive colonic mass located at the hepatic flexure. Biopsy results showed that the well circumscribed mass was positive for a histiocytic sarcoma. It was decided to start her chemotherapy as soon as her condition had improved.

After three months she underwent CT of her chest and her abdomen including the pelvic area. These scans showed bilateral pleural effusions and a pericardial effusion. In addition the scans showed an increase in the amount of nodules in the right lung (Figure 3).

Discussion

Histiocytic sarcoma is defined as a rare malignant form of cancer made up of histiocytes. Regardless of the site, this form of cancer tends to be problematic due to its aggressive nature and poor prognosis. It can occur in lymph nodes, skin and the gastro-intestinal tract^[1]. It may appear as a localized or disseminated form of cancer. Diagnosis is based on histological and immunohistochemical evidence of specific histiocytic lineage^[2].

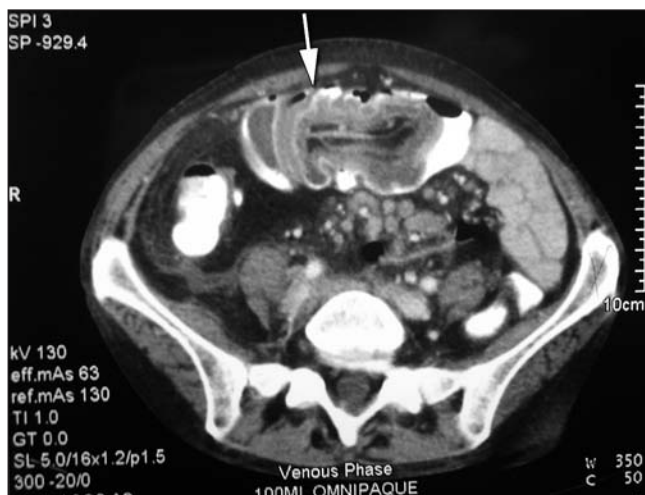


Figure 1: Abdominal axial CT image reveals bowel intussusception (see arrow).



Figure 2: Abdominal axial CT image reveals a large colonic mass located at the hepatic flexure (see arrow).

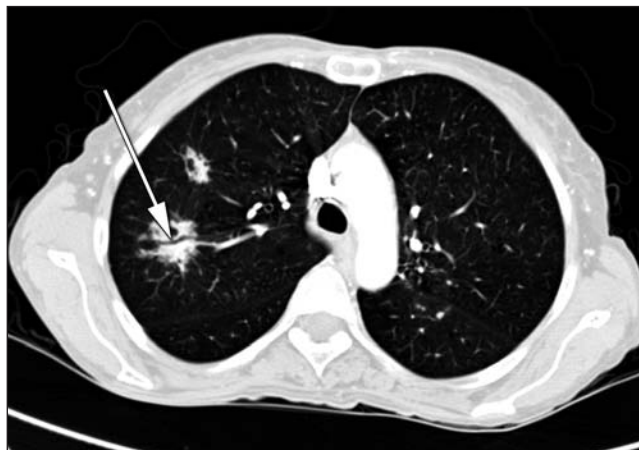


Figure 3: Chest axial CT image on lung windows reveals multiple spiculated lesions in the right lung (see arrow).

It is more commonly found in adults than children, with the average age being 46 years. It occurs equally in males and females. It accounts for less than 1% of all haematolymphoid neoplasms. One third present in the lymph nodes, 1/3rd in various other extra nodal sites, such as the intestinal tract, and 1/3rd in the skin [3]. Due to the rarity of this malignancy, survival rates are difficult to calculate. The size of the mass is said to be a predictor of survival. Masses equal to or greater than 3.5 centimetres have the worst outcome. Metastases can occur in lymph nodes, lung and bone [4]. In cases where the cancer presents as localized small masses there seems to be a positive long-term outcome [5].

The aetiology of histiocytic sarcoma is unknown and is thought to arise from antigen-processing phagocytes (histiocytes). It is a malignant creation of neoplastic cells that has immunophenotypic and morphologic features similar to histiocytes (tissue macrophages). Patients usually suffer from a fever, fatigue, weight loss, and weakness. Physical findings may include lymphadenopathy or hepato-splenomegaly and bones may show lytic lesions. Patients may also present with bowel obstruction [4]. The patient discussed in this report suffered from extensive lymphadenopathy and bowel obstruction. Such findings are in keeping with that described in literature.

Microscopically the tumour cells tend to be composed of sheets of large epithelioid cells and rich in eosinophilic cytoplasm. The cells may be focally spindled with oval or round nuclei ranging from mild to pleomorphic; large eosinophilic nucleoli and vesicular chromatin. The mass may be

well circumscribed or infiltrating in extra nodal cases. Evidence of histiocyte lineage is typically seen including expression of CD 4, CD 14, CD 31, CD 45 and CD 68. Lysozyme with a granular staining pattern is a definite sign of the presence of a histiocytic sarcoma. Markers of B-lineage (CD 20 and CD 79a), T-lineage (CD 3), myeloid (CD 33 and CD 34) and dendritic (CD 21, CD 35 and CD 1a) should be negative. Other negative markers include CD 30, CD 15 and CD 117 [6]. An important haemoglobin scavenger marker, CD 163, with a rare exception for identifying cancers of histiocytic lineage is another definite sign of the presence of a histiocytic sarcoma [2].

A diagnosis of a histiocytic sarcoma relies primarily on the presence of histiocytic lineage and the ruling out of other large cell malignancies. This is achieved by extensive immunophenotypic investigation. Immunohistochemistry also plays an important role in evaluating the differential diagnosis of histiocytic sarcoma [5].

The biopsy results of this patient were consistent with those of a histiocytic sarcoma. The specimen showed large, round cells with focal areas of spindling. Eosinophilic cytoplasm and pleomorphic basophilic nuclei were present.

The cancer progresses quickly particularly if the lymph nodes are affected. Histiocytic sarcoma has a poor prognosis due to its quick progression as well as the presence of widespread disease and its poor response to chemotherapy. Chemotherapy and radiation therapy can be used for local control of the malignancy [4]. Surgical removal of the malignancy is best suited for single small masses located in limbs [1]. Treatment of a histiocytic sarcoma ultimately

depends on the location, size and stage of the mass as well as the condition of the patient [3].

Conclusion

CT is the preferred imaging modality: it depicts the exact site of a malignancy and is useful to evaluate lymph node enlargement resulting from a malignancy; it is also preferred for following the progress of a gastro-intestinal malignancies [7].

The clinical course of a histiocytic sarcoma is often aggressive with an associated high mortality rate. Therefore, it is important to correctly recognise this rare malignancy using specific immunohistochemical markers because the correct diagnosis and early detection could lead to a better prognosis for the patient [6]. Awareness of this malignancy is important because masses can closely mimic other lymphoid tissue malignancies in their clinical presentation and morphologic appearance [3]. The patient under discussion was due to commence chemotherapy when her clinical condition improved.

References

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